

# Unusual Presentation of Sacral Spinal Dural Arteriovenous Fistula with Isolated Lower Limb Fasciculations and Restless Leg Syndrome

Dear Editor,

Spinal dural arteriovenous fistulas (SDAVF) are typically formed due to a pathological connection between spinal radicular artery and a radicular vein. They are the most common vascular malformations of the spinal cord and a treatable cause of progressive para or tetraplegia.<sup>[1]</sup> SDAVFs are acquired lesions with a diverse and often deceiving clinical presentation. Symptoms arise due to arterialization of a draining vein resulting in local venous congestion and cord edema.<sup>[2]</sup> SDAVFs more commonly occur in male patients in their 6<sup>th</sup> decade of life.<sup>[3,4]</sup> The vast majority of patients present with gradually progressive sensory and/or motor deficits, frequently associated with autonomic involvement. There is often a combination of both upper and lower motor neuron signs and symptoms. Here, we describe a case of SDAVF that involves an atypical clinical presentation.

A healthy male in his 60s presented to the neurointerventional clinic with a 6-month history of fasciculations involving both lower limbs, with the right leg more severely affected. The fasciculations were noted incidentally, and they were progressively increasing in frequency. There was no history of weakness or atrophy involving lower limbs. Additionally, he denied any bowel or bladder dysfunction, autonomic symptoms, or focal motor or sensory deficits in any other part of the body. His sleep hygiene worsened due to an uncomfortable sensation in both lower limbs, with a constant urge to move. He was diagnosed with restless leg syndrome (RLS) for which he received a pharmacological treatment without significant He eventually underwent imaging with a MRI spine, raising the possibility of a SDAVF.

On neurological examination, he was alert, oriented with normal language examination and cognition. His cranial nerves were intact without any tongue fasciculations or jaw jerk. Motor testing showed normal power in all four limbs. Both superficial and deep sensory examination was intact including perineal area sensory examination. Cerebellar testing was normal. Fasciculations were observed involving bilateral gastrocnemius muscles. There was no associated muscle atrophy. Reflexes were absent in both lower limbs and no pathological reflexes were elicited.

Nerve conduction (NCS) and electromyography (EMG) studies showed fibrillations and positive sharp waves in the lower limbs muscle groups along with low amplitude motor response consistent with axonal loss. A multimodal spinal MRI showed intradural flow voids between the roots of the cauda equina, extending from the sacral level to the conus medullaris without any T2 hyperintensities in the spinal cord. Contrast-enhanced

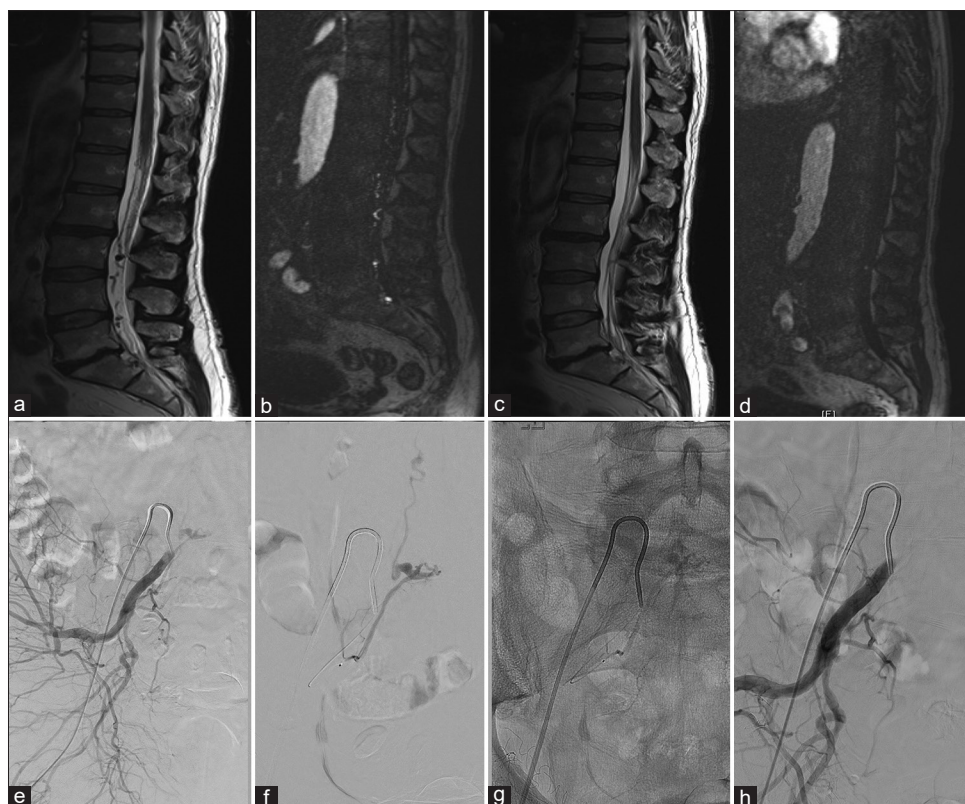
MRA spine showed enhancement of the flow voids with an arterial feeder to the fistula arising from the posterior division of the right internal iliac artery (Correlate with [Figure 1]). In retrospect, the SDAVF was reported on a previous MRI spine from 2011, initiated for lumbar back pain. Since he was otherwise asymptomatic at the time, there was no follow up of the MRI spine findings in 2011.

He underwent urgent spinal angiography and endovascular embolization under general anaesthesia. His spinal angiogram demonstrated a dural arteriovenous fistula, located in sacral region with the presence of dilated spinal perimedullary veins. The SDAVF had arterial supply from a hypertrophic iliolumbar artery arising from the posterior division of right internal iliac artery (correlate with [Figure 1]). Endovascular embolization was performed using 50% glue (NBCA, n-butyl cyanoacrylate) injected into the fistula and proximal part of the draining vein, with complete radiographic cure (correlate with [Figure 1]). Repeat spine MRI and contrast-enhanced MRA 3 months post-procedure, showed interval resolution of the previously noted flow voids (correlate with [Figure 1]). Clinically, his lower limb fasciculations have now stabilised without further progression. He also has near complete resolution of symptoms of RLS with improved sleep hygiene.

Overall, SDAVF are rare, frequently misdiagnosed spinal vascular malformations.<sup>[5]</sup> The most common location of fistula is lower thoracic or upper lumbar level. Initial symptoms of SDAVFs are often mild and non-specific, consisting of gait disturbances and sensory disturbances. Later, patients may develop worsening sensory and motor deficits, predominantly involving the lower limbs. Motor symptoms usually have a combination of both upper and lower motor neuron features. Bowel and bladder incontinence, erectile dysfunction are also common.<sup>[6]</sup> In contrast to cranial DAVFs, haemorrhage is extremely rare in SDAVFs.<sup>[3]</sup>

The presence of nonspecific symptoms that mimic common etiologies and the lack of pathognomonic features makes the diagnosis of SDAVFs challenging. The three characteristic findings of a SDAVF visualized in routine MRI spine are: spinal cord hyperintensities on T2-weighted images, the presence of flow voids in the intradural space, and spinal cord enhancement on T1 post-contrast.<sup>[2,7]</sup> However, spinal angiography remains the gold standard for the diagnosis. The goal of the treatment is complete radiographic cure with either microsurgical disconnection or endovascular embolization, alone or in combination.<sup>[8,9]</sup>

This case was illustrative from multiple aspects. Firstly, the clinical presentation in this case was unique and atypical



**Figure 1:** (a) Pre-treatment, Sagittal T2 image showing flow voids at the cauda equina region (b) Pre-treatment, post-contrast Sagittal T1 image showing enhancement of flow voids (c) Post-treatment, Sagittal T2 image showing resolution of flow voids (d) Post-treatment, post-contrast Sagittal T1 image showing resolution of flow voids (e) Right internal iliac artery angiogram showing SDAVF (f) Microcatheter run of lliolumbar artery (g) Native image of the microcatheter run (h) Post-treatment right internal iliac artery angiogram showing complete occlusion of SDAVF

of a SDAVF. The patient presented with fasciculations in the absence of any other motor deficits, and this has not been described previously in the literature.<sup>[2,6]</sup> Also, this patient presented with RLS-like sensory symptoms that subsided after endovascular cure of the SDAVF. This RLS-like presentation has not been described in association with SDAVFs.<sup>[10]</sup> The pathogenesis and clinical presentation of SDAVFs results from AV shunting causing chronic venous hypertension and local tissue hypoxia and progressive myelopathy. The differences in segmental venous outflow pattern contributes to the phenomenon whereby venous hypertension and congestion is transmitted in a caudo-cranial and postero-anterior direction independent of the anatomical location of the fistulous connection. Our patient had limited venous drainage of the anterior compartment, that resulted in an unusual earlier involvement of the motor neurons with fasciculation and milder sensory symptoms in the form of RLS-like presentation. Prior to his treatment, our patient had a rapidly progressive RLS that significantly affected his sleep despite being on medications. There is a clear temporal correlation between stabilization of motor symptoms as well as an improvement in sensory symptoms with endovascular treatment of SDAVF. These factors are highly suggestive that the fasciculations and RLS were the result of his underlying SDAVF. The prolonged

asymptomatic course of spinal dural fistula is very unusual and described more commonly in cervical lesions. The recent symptomatic progression of disease may be related to the dynamic nature of SDAVF with deterioration in function of radicular veins over time.<sup>[2]</sup> The sacral location of fistula is uncommon and described in only 4% cases and often better suited for endovascular management.<sup>[1]</sup> This patient also did not have any T2 hyperintensities within the spinal cord on MRI, however, this has been described in up to 6.4% cases.<sup>[3]</sup> One of the limitations of the case is lack of follow up nerve conduction and electromyography studies for correlation. We did advised for repeat electrodiagnostic testing, however, the patient declined. Most importantly stabilization of his motor features and improvement in clinical symptoms after endovascular cure, has a higher likelihood to exclude other etiologies as the underlying cause of his clinical presentation.

#### Financial support and sponsorship

Nil.

#### Conflicts of interest

There are no conflicts of interest.

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**Submitted:** 27-Dec-2020 **Revised:** 06-Jan-2021 **Accepted:** 12-Mar-2021

**Published:** 03-Jun-2021

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**DOI:** 10.4103/aian.AIAN\_1245\_20